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Case Report Open Access

The Role of 3-Dimensional Ultrasound for the Diagnosis of LPSVC and A Review of the Literature

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Abstract

Objective: The purpose of this article is to review the published literature and determine the rolethat3-dimensional Ultrasonography (3DUS) play in the diagnosis of LPSVC.

Methods: The volume data sets were acquired with transverse sweeps through the fetal chest. Volumes were stored in the ultrasound machine's hard drive and subsequently analyzed offline.

Results: PLSVC was unequivocally diagnosed and the diagnosis confirmed by postnatal US.

Conclusion: 3D ultrasonography especially power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus. The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30 -week fetus demonstrates the added value of 3D ultrasonography and especially 3D power colour in fetal echocardiography.

Case Presentation

A 39-year-old woman, gravida 2 para 1, presented at 30 weeks' gestation for detailed targeted organ scanning with fetal echocardiography, after having received contradictory opinions concerning a suspected atrioventricular canal defect during previous ultrasound examinations. Karyotype, which had been tested because of advanced maternal age, was normal. Targeted organ scanning for exclusion of fetal anomalies was performed, including complete fetal echocardiography 2 and 3 D US and coulor Doppler and 3 D Power Doppler power imaging (Figures 1-3).

In the present case, the initial diagnosis of LPSVC was made on observation of a dilated coronary sinus and an extraneous vessel identified to the left of the ductal arch in the three-vessels and trachea (3VT) view of the fetal heart (Figures 1E ,F and 3D).

We performed ultrasound examinations using a GE Voluson 730 pro. (Voluson 730, GE Healthcare, Milwaukee, WI, USA) using a 3-5-MHz mechanical volume transabdominal transducer). The volumes were stored and subsequently analyzed offline.

Meticulous inspection of the fetal anatomy did not reveal any associated cardiac or extra cardiac anomalies and fetal growth was on the 50th centile. A male neonate was spontaneously delivered at38 weeks of gestation with a birth weight of 3150 g with Apgar scores of 8, 9 and 10 at 1,5 and 10 min, respectively. Postnatal adaptation was uneventful. Postnatal echocardiography confirmed the prenatal diagnosis.

3D power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus (Figures 1 and 3). The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30-week fetus demonstrates the added value of power colour Doppler 3D in fetal echocardiography.

It appears that 3Dpower Doppler imaging applications will make a significant contribution to our understanding of the developing fetal heart in both normal and anomalous cases, to interdisciplinary management team consultation, to parental counselling, and to professional training

Comment

Persistent left superior vena cava is a rare but is the most common congenital anomaly of the systemic thoracic venous return. It is found in 0.3-0.5% of the general population [1-7]. The reported prevalence of persistent in CHD patients varies widely according to the diagnostic tool utilized it ranges from 1.3%, 2%, 3 to 10% and in up to 12% in association with congenital heart diseases [2,5,7-8].

In normal fetal development, the paired anterior cardinal veins drain the upper body and extremities of the fetus and become connected by an oblique vessel. During the 8th week of embryological development, a communication forms between the veins (future innominate [brachiocephalic] vein). The distal end of the left anterior cardinal vein degenerates (forms the ligament of Marshall), and is represented at birth by a small vestigial vein adjacent to the posterior wall of the left atria (Marshall's oblique vein) and by a fibrous band attached to this vein (Marshall's ligament), the remaining right anterior cardinal vein ultimately becomes the Right Superior Vena Cava (RSVC).

The distal end of the left anterior cardinal vein drains into the left horn of the sinus venosus (future coronary sinus) before it degenerates; this explains the drainage pattern of a PLSVC into the coronary sinus Failure of closure of the left anterior cardinal vein during cardiac development results in PLSVC [1,4,9-11].

Failure of the distal left anterior cardinal vein to degenerate is possible with or without degeneration of the right cardinal veins (Figure 3A). A genetic culprit may be genes for left- right signalling [12]. In general, Patients with left superior vena cava (LSVC) usually

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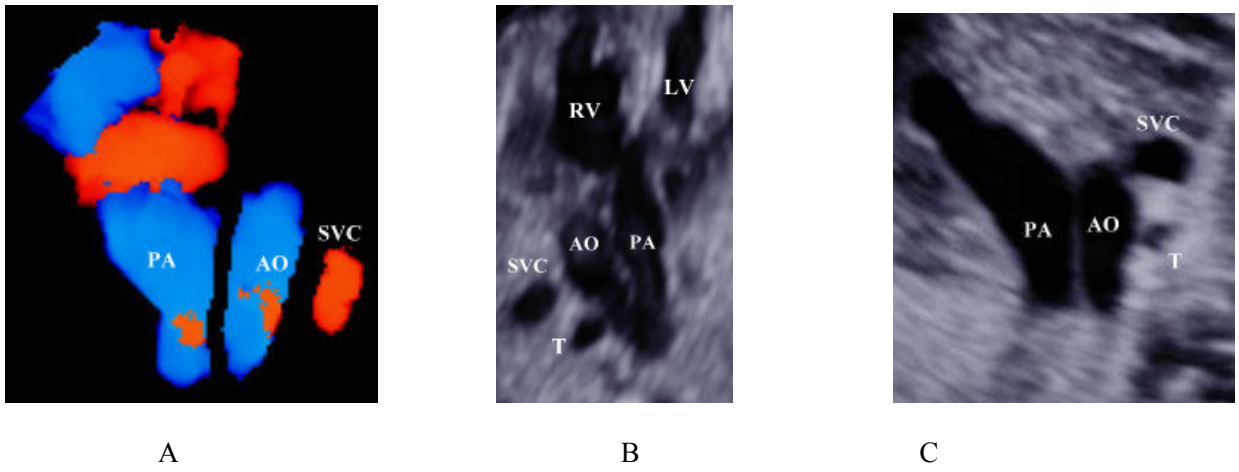

Figure 1: Transverse view of the upper mediastinum A, B, C: Normal three-vessel view. The 3-V V and trachea.
RA, right atrium;Left ventricle , right common carotid artery; RV, right ventricle; SVC, superior vena cava; T, trachea; Ao , aorta , PA, pulmonary artery.

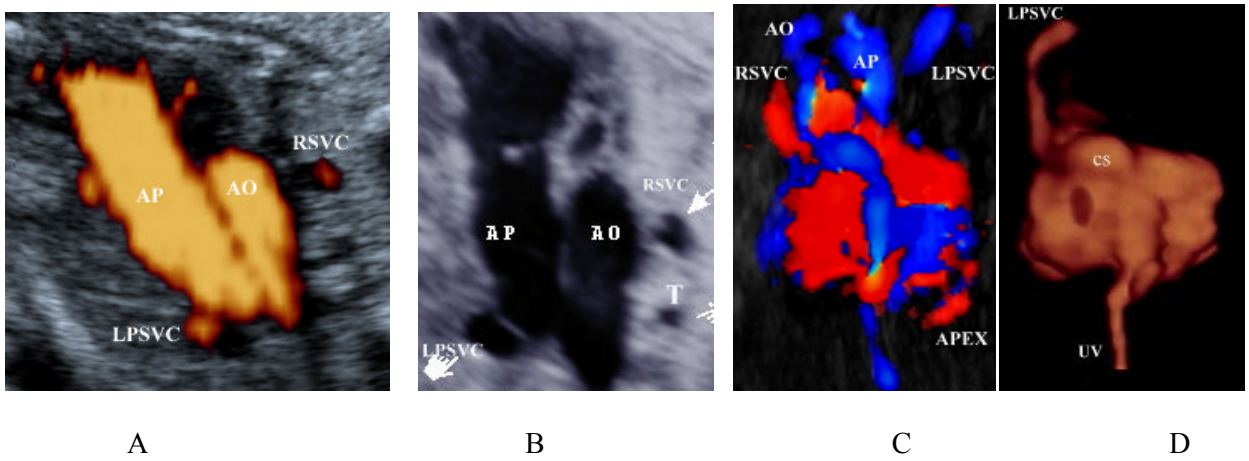

Figure 2: A, B and C: Four vessel view showing a left persistent superior vena cave (LPSVC).
D: 3D power Doppler ultrasound showing the LPSVC drains into the dilated coronary sinus and to the right atrium.
RA, right atrium;Left ventricle , right common carotid artery; RV, right ventricle; SVC, superior vena cava; T, trachea; Ao , aorta , PA, pulmonary artery , RSVC, right superior vena cava , PLSVC, persistent left superior vena cava , RA, right atrium , UV, umbilical vein , CS, coronary sinus.

have a normal right superior vena cava are asymptomatic, and the condition is infrequently detected a casual finding during an imaging test (CAT, NMR or venography) or during intervention (catheterization, pacemaker or defibrillator implantation, or surgery), and it has only practical implications when considering a left superior approach for implantation of a pacemaker, a cardioverter-defibrillator, or positioning of a central venous line [13,14].

When isolated, LPSVC usually has no clinical significance; however, it has been reported to occur in isolation in only 9% of cases [15]. Associated congenital cardiac defects (atrial septal defects, ventricular septal defects, endocardial cushion defects, and tetralogy of Fallot) and high incidence of rhythm disturbances may be detected in patients with PLSVC [16]. Presence of a LPSVC was first reported by Le Chat in 1787, and the first account of foetal structures comprising LPSVC was published by Marshall in 1850 [17].

According to Schummer, the superior vena cava system can be classified as type I, normal anatomy; type II, only LPSVC; type IIIa, right and left superior vena cava with connection; and type IIIb, right and left superior vena cava without connection [18,19]. The most frequent variant (82%) of the normal anatomy is the simultaneous presence of both right and left cava veins (double superior vena cava, type III) and so this anomaly is often missed as central line catheters are more commonly inserted on the right side [20,21]. Persistence of Left SVC with, absent right superior vena cava (type II) occurs in only 0.09-0.13% of patients with congenital heart disease [11]. It is often an isolated anomaly [21].

Left SVC drains either into right atrium via coronary sinus or into left atrium. 92% of Left SVC drains into right atrium via left portion of sinus venous or coronary sinus and are asymptomatic [22]. They are considered to be anomaly of coronary sinus [23]. In this case, the ostium of the coronary sinus opens directly into the right atrium in close proximity to the insertion of the atrioventricular valves [24]. If the coronary sinus is dilated, it can create the appearance of an atrioventricular canal defect (as in the present case) Park et al presented three cases in which the correct diagnosis avoided an unnecessary termination of pregnancy [25]. In 8% of cases, it connects to the left atrium in such variants with absent or unroofed coronary sinus or normal coronary sinus and so creates a right-to-left shunt and

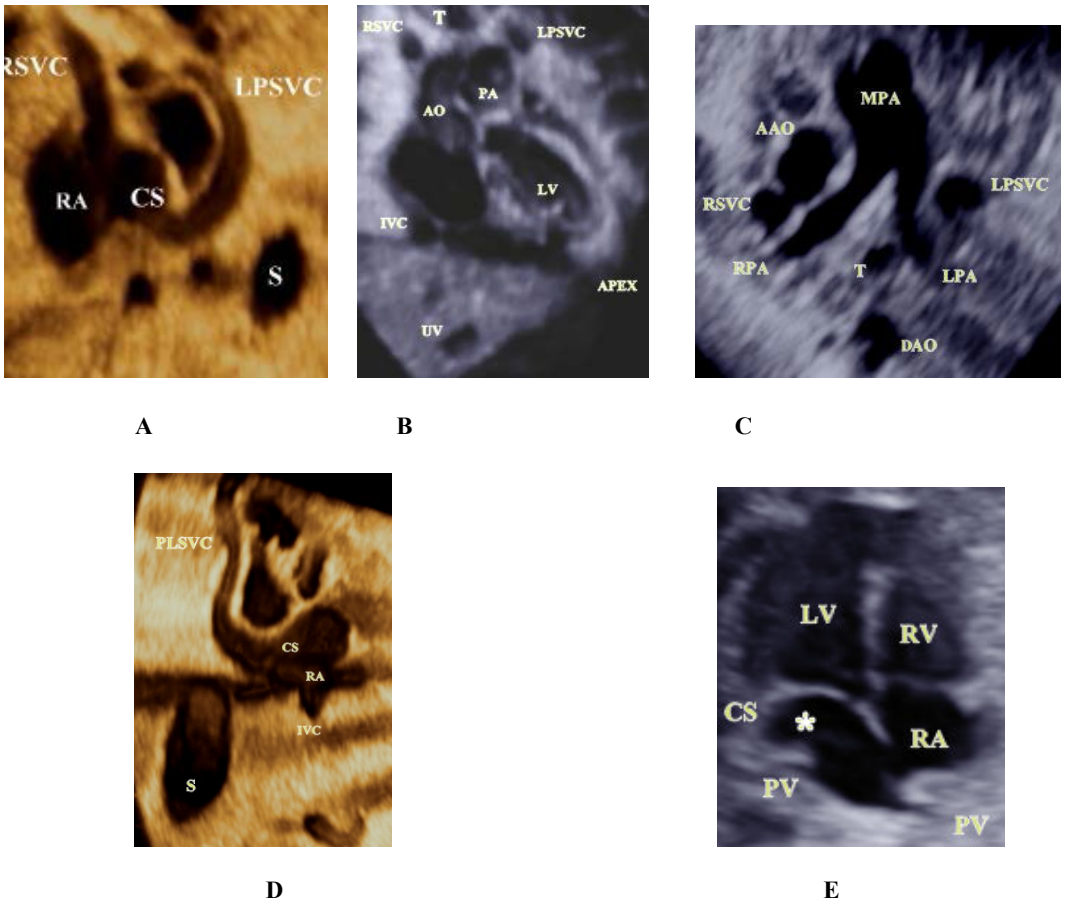

Figure 3: Volume-rendered images A, B, C: showing 2 superior vena cavas, one on the right and the other on the left side.
D-Oblique parasagittal ultrasound image of the patent left superior vena cava (LSVC), the dilated coronary sinus (CS) and the right atrium (RA).
E-4 chamber view showing dilated coronary sinus (star).
AO: aorte , vein , RA: right atrium , LPSVC: left persistent superior vena cava , RSVC : right superior vena cava , CS: coronary sinus , UV: umbilical vein , IVC : inferior vena cava.
AAO ascending aorta, MPA main pulmonary artery, MV mitral valve, LPV , left pulmonary vein, RPV, right pulmonary vein , SVC superior vena cava, TV tricuspid valve , Asterisks denote dilated coronary siuns.

paradoxical emboli and [26,27]. 3D power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus (Figures 1 and 3). The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30 -week fetus demonstrates the added value of power color Doppler 3D in fetal echocardiography.

In addition, 3D/4D image processing is advantageous compared with 2D ultrasound because detailed offline evaluation is possible; spatial relationships between lesions are easily demonstrated; blood vessel courses can be followed completely; volume calculations provide more accurate information on areas; and volumes can be inspected from different angles [28]. The digital storage capabilities of 3D/4D imaging enable remote diagnosis. The virtual volume can be stored and reloaded to allow follow-up examinations, comparisons, video conferencing, or revision of initial diagnosis. Furthermore, the volume dataset can be analyzed offline or reviewed by experts in another centre connected by the internet [29].

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